AMENDMENTS TO THE SPECIFICATION:

Please insert the attached "Sequence Listing" as separately numbered pages 1 - 8 after the abstract.

Please replace paragraph 6, with the following rewritten paragraph:

[006] Fig. 1: Protein domains analyzed for K8/K18 mutations and an example of the identification of a K8 mutation. (A) A central rod domain, consisting of α-helical subdomains, is flanked by non- α-helical head/tail domains. The head/tail domains are further subdivided into E, V and H regions. The subdomains of the rod are connected by nonhelical linker (L1, L1-2, L2) regions. The amino acid (aa) regions in black bars represent 5 domains that were examined for K8/K18 mutations. The remaining regions in gray bars contain 10 exonic K8/K18 domains that have been analyzed for mutations. (B) PCR products, from a control patient (with K8 WT) and a patient with K8 R340H, were analyzed by denaturing HPLC using a WAVE® System. The control (K8 WT; SEQ ID NO:5, SEQ ID NO:6) is characterized by one major peak, while the K8 R340H; SEQ ID NO:7, SEQ ID NO:8 shows a different chromatogram due to resolution of the homoduplexes from the heteroduplexes, thereby suggesting the presence of a K8 mutation. Electropherograms from DNA sequencing confirm the presence of a K8 R340H heterozygous missense mutation (CGT→CAT).

Please replace paragraph 8 with the following rewritten paragraph:

[008] Fig. 3: K8 R340H mutation-proximal comparison of type II keratin sequences and confirmation of K8 R340H mutant protein expression in explanted livers. (A) (SEQ ID NO:9) Single letter abbreviations are used to represent amino acids. Bold dots represent amino acids that are identical to the K8 sequence. The shaded area highlights the conserved R340 of K8 and shows the histidine mutation we identified. Note that the K8 R340-containing motif (AEQ***R***G; SEQ ID NO:9, residues 4-8) is highly conserved in type II keratins. It is also conserved across species, being found in mouse and frog K8. (B) BHK cells were transiently cotransfected with K8/K18 WT or K8 R340H/K18 WT. K8/K18 immunoprecipitates were obtained from 1% NP40-solubilized cell lysates. The immunoprecipitates were analyzed by SDS-PAGE, followed by immunoblotting with anti-K8 R340 or anti-K8 H340 epitope-specific antibodies that preferentially recognized K8 WT or K8 R340H mutant, respectively. (C) K8/K18 immunoprecipitates were obtained from 1% NP40-solubilized normal liver or livers with the K8

R340H mutation. Samples were analyzed as described in panel B. Note that anti-K8 R340 (WT) recognizes K8 in the control patient (with K8 WT) and the patient with K8 R340H (lanes 1-6), whereas anti-K8 H340 (mutant) recognizes only patient livers with the K8 R340H mutation (lanes 2-6) but not the normal liver (lane 1). This indicates that the patients with K8 R340H mutation are heterozygous with regard to the keratin mutation. Arrowheads correspond to degraded K8.